Hardaway (W. A.)

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## MULTIPLE XANTHOMA,

EXHIBITING THE PLANE, TUBERCULAR AND TUBEROSE VARIETIES OF THE DISEASE; WITH REMARKS.

By W. A. Hardaway, M. D., Prof. Dermatology in the Mo. Med. Coll.; Consulting Dermatologist to the City and Female Hospitals of St. Louis.

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## A CASE OF MULTIPLE XANTHOMA, EXHIBITING THE PLANE, TUBERCULAR AND TUBEROSE VARIETIES OF THE DISEASE; WITH REMARKS.

By W. A. HARDAWAY, M. D., Prof. Dermatology in the Mo. Med. Coll.; Consulting Dermatologist to the City and Female Hospitals of St. Louis.

[ Read Before the Am. Dermatological Association, West Point, New York, August 27, 1884.]

M., æt. 44, born in Germany, occupation cook, was . sent to me for examination by Drs. Dorsett and

Epstein, of the St. Louis Dispensary.

It seems that in 1876 he was a patient in the City Hospital. and for the notes of his case at that time, and for his early history, and also for opportunities of seeing the patient after his readmission to the hospital, I am indebted to Dr. Dean, the superintendent and physician in charge. His father and mother were healthy people and lived to an advanced age. He himself had been strong and well, except having had ague, up to four years before his admission to the hospital. From his boyhood he had suffered from hyperidrosis of the feet to an excessive degree. He also perspired freely from the general surface. While he was working before an intense fire every day from 3 to 11 o'clock, he began to stop sweating, and in its stead he experienced severe burning and itching, and two years after it was noticed that he was turning yellow. During this time his sleep and appetite were poor. His food frequently regurgitated. He would sit by an open window to ease the burning and itching of his skin.

When Dr. Dean saw the patient, his right side was almost of a bronze color, but the left side was yellow; heart and lungs sound; liver enlarged. He had been treated in the hospital since December 18, 1876 for chronic hepatitis, and left June 12, 1876. He remained out of the hospital for a number of years. Happening to apply to the City Dispensary for relief on May 23, 1884, he was referred to me, and shortly afterwards was again admitted to the City Hospital. At this time the diagnosis of xanthoma was made, and I had the pleasure of showing the case to the members of the St. Louis Medico-Chirurgical Society. The following notes were then taken:

According to his best recollection it is now four years ago that he first noticed xanthomatous lesions in the gluteal region. The various growths appeared gradually, but in what order he does not know. From the very beginning of the discoloration of his skin, twelve years since, up to to-day, he has suffered from an agonizing pruritus of the general surface, and it is for this that he has sought relief at the hands of physicians. He says that he has never experienced headache in his life.

Present Condition.—Patient is about 5 feet 7 inches in height, and weighs 138 pounds. His appetite is ravenous, but at the same time he says he feels quite weak, and occasionally has attacks of dizziness. Pulse is feeble. Has emphysema of lungs, with consequent asthmatic paroxysms. Temperature is normal. He passes about 65 ounces of urine daily, which is of a dark amber color, of acid reaction, specific gravity 1016, and shows a trace of sugar. No albumen or casts. Stools, elay color. The liver is enlarged laterally, much thickened and apparently nodular; has well defined edge. The dullness begins one-half inch below nipple, and extends to three inches below the level of the ribs, and two inches to left of the epigastrium.

A general inspection of the patient reveals the following: He is evidently feeble, and looks quite decrepit, and has a stooping gait and shuffling walk. The color of his skin at once strikes the attention. It is of a decided bronze hue, almost black, and darker in some situations than in others. The conjunctivæ are also involved in the general discoloration, but are much lighter than the skin. Indeed, the man looks as if he were the subject of Addison's disease rather than jaundice. The whole integument shows the characteristic markings of an intense and uncontrollable pruritus—it is torn, thickened and scarred, and the hairs on the scalp and elsewhere on the body have been almost scratched away.

Face and Head.—On the upper lids are to be noted the characteristic patches of xanthoma planum. They are of a much lighter color than the surrounding skin, and are all the more conspicuous on that account. They extend, on each upper lid, from one canthus to the other, and, perhaps as the result of coalescence, occupy nearly the whole width of the lid. They are elevated above the level of the skin, but possess no decidedly marked tuberculated edges. On the upper lids one may see at regular intervals closely packed black dots, the apertures of gland ducts. On the lower lids, and extending somewhat down on the cheeks, the patches are smooth, irregular, and on a level with the skin. On the upper lip are four tubercles from pinhead to pea size. Scattered over chin are a number of small tubercles. On the left ear are four tubercles, and on the right, five, pea-sized and apparently involving the whole thickness of the skin.

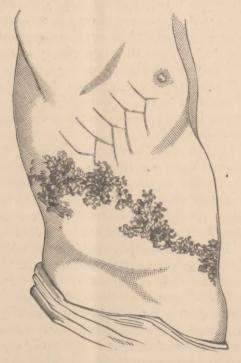
Upper Extremities.—Over the right olecranon process is a double tumor, each half the size of a pigeon's egg, and over the region of the elbow generally there are about twenty-five tubercles varying from a pinhead to a pea in size. Over the left olecranon are two aggregated masses of the bigness of a hazel-nut, and in the same region are eight or ten smaller tubercles. On the outer border of the right wrist is a hazel-nut sized growth, and on the back of the wrist is also a small tumor. On the right index finger, at the metacarpo-phalangeal junction, dorsal surface, is a tumor the size of a hazel-nut. Over the first joint of

index and second fingers are two tumors on each of the size just mentioned. On the last phalangeal joints of first, second, third and fourth fingers are growths somewhat smaller than a hickory nut.

On inspecting the palmar surface of the right hand, we find an infiltrated yellow line exactly occupying the furrow which divides the ball of the thumb from the palm. In the crease over the first phalanx of the thumb is a congeries of flat vellow bands. The whole ball of the thumb is a mass of closely packed tubercles. The index finger, palmar surface, of this hand, has a broad band running across the second phalanx. On the palmar surface of second finger is a solid mass of tubercles, some more prominent than others, and here and there of a yellower color, extending from tip of finger to metacarpo-phalangeal junction. This same condition is also to be observed on the palmar surfaces of third and fourth fingers. Upon scrutiny of the dorsal surface of the left hand the xanthomatous growths are found to be disposed as follows: On the inner border of the wrist are three tumors, two large and one somewhat smaller. The small one is the size of a pea, and the others, as on the other hand, are as large as hazel-nuts. On the second joint of the first finger is a filbert-sized growth. On the second finger is a small tubercle over the metacarpo-phalangeal joint, and on the second phalangeal joint is a hazel-nut sized tumor. The joint is decidedly enlarged at the metacarpo-phalangeal articulation of the second finger. A tubercle is to be distinctly felt on the tendon of the extensor indicis. On the inner border of the second finger near the second phalangeal joint is a large mass of tumors of the size of a half of a hickory nut. Over the last joint of this finger are two growths the size of a pea and one as big as a hazel-nut. On the third finger, second joint, a hazel-nut sized patch and over the last joint are two-one the size of a hazelnut and one the size of a pea. Over the little finger, last joint, is one growth, the size of a hazel-nut. Over the left elbow are two large tumors, the size of a pigeon's egg, and a number of smaller ones in the vicinity.

Trunk. Commencing at the spine, and extending between the tenth and twelfth ribs, and obliquely upwards to the ninth

rib, then crossing the ninth and tenth ribs on a line dropped from the axilla to the crest of the ilium, and then obliquely downward to the umbilicus, and strictly limited by the median line, are observable clusters of innumerable yellow xanthomatous tubercles, having the exact arrangement of herpes zoster. These clusters of tubercles form a band on the right side of the



body two inches in width, and exactly limited by the spine in the rear and the median line in front. In three or four places a dozen or more tubercles have coalesced, making small quarter dollar sized plaques.

The other hundreds of tubercles, although touching each other on all sides, still retain their individuality. They are slightly elevated above the line of the skin, and in many instances are clearly umbilicated. From their corymbose grouping and other features it is manifest that they correspond to the

distribution of the cutaneous nerves of the region involved. These tubercles date from an early period of his disease. He gives an indefinite account of neuralgic pain in these parts before the growths appeared. (See illustration).

Over the left gluteal region near the cleft, is a group of nineteen tumors, ranging from a split-pea to a hickory nut in size, and two inches below, just above the gluteal fold are three large hickory nut sized tumors. In the cleft between the nates is a band-shaped growth made up of distinct tubercles. Just above gluteal fold on right side is a cluster of twenty tumors as big as hickory nuts, and scattered over buttocks are eight the size of a split pea. On the back of the thigh of each leg are three small, pea-sized lesions. On anterior surface of the thighs are a few—some twelve on each—of the same dimensions, scattered up and down the parts.

Over the right knee are four tumors, ranging from split pea to hazel-nut, and over the left knee is one the size of a hazel-nut. On the tibia of each leg, just below the attachment of the lig. patellæ, is to be seen and felt a large node of the size of half of a hen's egg. Over the left heel, internal surface, is a xanthomatous lesion the size of a half hen's egg, and two the size of hazel-nuts. In and on the tendo Achillis is a large growth the size of a pigeon's egg, and two the size of peas.

On the external surface of the heel is one flat infiltration the size of a half dollar, and two the size of pigeons' eggs. Under the os calcis are eleven flat, yellow infiltrations from the size of a silver three cent piece to that of a silver dime. On the internal border of foot is an aggregated patch as big as a quarter dollar. On external border of foot (left) about the middle, is a reddish-yellowish tumor half the size of a hen's egg. Over the metatarso-phalangeal joint is an infiltration the size of a half dollar. Over inner side of great toe is a yellowish patch the size of a half dollar. Over crest of tibia of the right leg is a number of tumors the size of half of a hen's egg. These growths do not rise prominently above the level of the skin, but are quite appreciable to the touch. On the inner side of calf one lesion, the size of a hazel nut. On tendo-Achillis of this leg also are several tumors, one the size of

a small hen's egg. On external and internal surface of heel are two aggregations, each the size of a half dollar, made up of small tubercles. There is a large tumor on the outer border of this foot about the size of, and in the same situation as that on the left foot. On the second toe are four small tubercles and also on the dorsum of the foot are a few scattered tubercles. The internal and external malleoli are enlarged to the size of half a hen's egg, and stand out prominently. On the glans penis is a a single intensely yellow tubercle the size of a pea, and on the scrotum are a dozen similarly sized very yellow lesions. There are no growths on the scalp. On the chin a few of the tubercles are pierced by hairs.

At my request Dr. J. C. Mulhall kindly made an examination of the patient's mouth and larynx, and sent me the following report:

May 24, 1884.

DEAR DOCTOR—The man Meyer presented the following appearances:

At the lower sulcus, between jaw and cheek, a continuous irregular yellow staining (the upper sulcus uninvolved). A like appearance involving the soft palate, stopping definitely at its junction with the hard portion. The vocal cords are of a pale yellow hue. In the trachea, anteriorly, from the lower border of the cricoid to the second tracheal ring are several pinhead infiltrations, and just at the lower border of the cricoid anteriorly, two large flat infiltrations with irregular borders, each slightly larger than a kidney bean, and symmetrical in every particular.

Their situation precludes any possibility of being a factor in the dyspnea of which the man complains.

Yours truly,

J. C. MULHALL.

Color of Lesions.—The color of the plaques on the eyelids presents the fawn or buff tint usual in this situation. The tubercular and tuberose growths vary in tint. Most of the small isolated tubercles are yellow; in some places, where they have massed together the masses are bronzed like the skin. Many of

the tumors are a darkish red, but when pressed upon become yellow, not as a whole, however, but in a mottled way.

Symptoms. The general symptoms are mostly referable to the patient's emphysema, and to a marked distention of the abdomen. There is also tenderness and occasional sharp pains over the hepatic region. I have already mentioned his pruritus. The lesions themselves give him no inconvenience, except in a mechanical way.

When pressed upon firmly no particular sensation is experienced, but if he should knock the large tumors on his hands against some article, there is aroused some smarting and burning. There certainly is no spontaneous pain, nor pain on direct, firm pressure. The various lesions, however, participate in the general pruritus.

I may state in this place that a few of the tumors over the knuckles are quite gristly, but in other situations, even the large masses are soft. I could not determine that the plane variety ever began as tubercles, or that extension took place by the deposit and subsequent fusion of papules. On the lids, the patches seemed to grow by peripheral extension. In the centres of many of the tubercles on the side were to be observed minute black dots, like plugged orifices of glands.

Remarks.—In making a general review of the case, the following points may be noted:

- 1. It is established that the bronzed hue of the skin preceded the xanthoma by a number of years.
- 2. The eruption is perfectly symmetrical, with the exception of the curiously arranged lesions on the side of the trunk.
- 3. The involvement of the mucous membranes, and the undoubted implication of the internal organs.
- 4. The peculiar configuration and location of the tubercles on the side, showing the probable influence of the nervous system in the evolution of xanthomatous lesions.
- 5. The presence of xanthomatous growths on tendons and in the areolar tissue, but more particularly the probable implication of bone structure in the same process.<sup>1</sup>

<sup>1</sup> T. C. Fox (Lancet Nov. 9, 1879) reports a case which he in another

- 6. The evidence, though slight, of sugar in the urine.1
- 7. The patient states that some of the growths have disappeared; but of this fact I have no personal knowledge—it is clear, however, that new lesions are still constantly developing.

From a careful consideration of this and other cases that have come under my observation, and a thorough study of the literature of the subject, I am inclined to suggest that xanthoma is a diathetic affection, and that its connection with the liver, and the frequent jaundice, occurs only in a secondary way. In other words, it seems to me plausible, that when jaundice precedes the xanthoma it is because xanthomatous lesions have been primarily deposited in the liver; and unless this occur, there may just as well be a xanthoma without jaundice—as has, in fact, been often seen; but that the liver is peculiarly prone to these growths, and that it is for this reason we see so many cases of xanthoma which have been preceded by, or associated with, jaundice. A study of the cases of hereditary xanthoma<sup>2</sup> and of

place (Epitome of Skin Diseases. Phila., 1883) speaks of as presenting "remarkable bone changes of a gouty or rheumatic character." On examination of the original report (lov, cit.) we find that the patient was a markedly rheumatic and gouty subject, and that the bone changes in question consisted of pain and swelling back of the right os calcis, and swellings about the phalangeal and metacarpo-phalangeal joint. Certainly this state cannot be considered as strictly analogous to the nodes and enlargements observed in my case. Then again, my patient gave no rheumatic or gouty history.

I Hutchinson says that the two cases of vitiligo tuberosa which have been recorded in association with diabetes, in which the eruption came out suddenly, and also showed a tendency to cure, were perhaps examples of a different malady. (Hebra and Kaposi Syd. Soc. edit, p. 354) Mr. M. Morris has described an eruption which he says seems allied to xanthoma, which was observed in connection with diabetes. The differential diagnosis seemed to be founded on the absence of jaundice, the solidity of the lesions, the freedom of the lids, the presence of the spots in the neighborhood of hair follicles, and the absence of complete yellow coloration in the lesions. In this present case, tubercles were pierced by hairs, some of the nodules on the fingers were hard, and some lesions were brown.

<sup>2</sup> For cases of hereditary xanthoma see reports by Barlen, Brit. Med Jour., May 24, 1884, p. 998; and Eichoff, Deutsche Med. Wochinscher., 1884, No. 4. Church, St. Bartholomaw's Hosp. Rep., 1874, vol. x., quoted by Legg.

the numerous cases in which jaundice was absent only lends strength to this view. It is also probable that in persons having the xanthomatous diathesis, certain conditions of irritation, or unusual motion and the like, may provoke a local deposit in situ; for instance, in the lids, over joints, etc. Thus my patient may have had his peculiar zosteriform deposits evoked by an abortive zona.

For much assistance in taking the notes of the preceding case, I am indebted to my friend, Dr. E. W. Hodsden.





